

**AMENDMENTS TO THE CLAIMS**

1. (Original) An animal having a heterologous nucleic acid sequence replacing an allele of an *atonal*-associated nucleic acid sequence under conditions wherein said heterologous sequence inactivates said allele.
2. (Original) The animal of claim 1 wherein said heterologous nucleic acid sequence is expressed under the control of an *atonal*-associated regulatory sequence.
3. (Original) The animal of claim 1 wherein both *atonal*-associated alleles are replaced.
4. (Original) The animal of claim 3 wherein said heterologous nucleic acid sequences are nonidentical.
5. (Original) The animal of claim 3 wherein said animal has a detectable condition.
6. (Original) The animal of claim 5 wherein said detectable condition is selected from the group consisting of loss of hair cells, cerebellar granule neuron deficiencies, hearing impairment, an imbalance disorder, joint disease, osteoarthritis and abnormal proliferation of cells.
7. (Original) The animal of claim 1 or 3 wherein said heterologous nucleic acid sequence is a reporter sequence.
8. (Original) The animal of claim 1 or 3, wherein said *atonal*-associated allele is replaced with an *atonal*-associated nucleic acid sequence under the control of regulatable promoter sequence.
9. (Original) The animal of claim 1 or 3, wherein said *atonal*-associated allele is replaced with an *atonal*-associated nucleic acid sequence under the control of a tissue-specific promoter sequence.
10. (Original) The animal of claim 1 or 3 wherein said animal is selected from the group consisting of a mouse, *Drosophila*, zebrafish, frog, rat, hamster and guinea pig.

11. (Original) A method for screening for a compound in an animal wherein said compound affects expression of an *atonal*-associated nucleic acid sequence comprising:

delivering said compound to said animal, wherein said animal has at least one allele of an *atonal*-associated nucleic acid sequence inactivated by insertion of a heterologous nucleic acid sequence, wherein said heterologous nucleic acid sequence is under control of an *atonal*-associated regulator sequence; and

monitoring for a change in said expression of said *atonal*-associated nucleic acid sequence.

12. (Original) The method of claim 11 wherein said compound affects expression of an *atonal*-associated nucleic acid sequence.

13. (Original) The method of claim 11 wherein said compound affects a detectable condition in an animal.

14. (Original) The method of claim 11, wherein said heterologous nucleic acid is a reporter sequence.

15. (Original) A method for screening for a compound in an animal, wherein said compound affects a detectable condition in said animal, comprising:

delivering said compound to said animal wherein at least one allele of an *atonal*-associated nucleic acid sequence in said animal is inactivated by insertion of a heterologous nucleic acid sequence, wherein said heterologous nucleic acid sequence is under the control of an *atonal*-associated regulatory sequence, and monitoring said animal for a change in the detectable condition.

16. (Original) The method of claim 11 or 15 wherein said compound affects said detectable condition.

17. (Original) The method of claim 15 wherein said compound affects expression of said heterologous nucleic acid sequence.

18. (Original) A method of treating an animal with a deficiency in cerebellar granule neurons or their precursors comprising delivery of a therapeutically effective amount of an *atonal*-associated amino acid sequence or nucleic acid sequence to a cell of said animal.

19. (Original) A method of promoting mechanoreceptive cell growth in an animal, comprising delivering a therapeutically effective amount of an *atonal*-associated amino acid sequence or nucleic acid sequence to a cell of said animal.

20. (Cancel)

21. (Original) A method of treating an animal for hearing impairment, comprising delivering a therapeutically effective amount of an *atonal*-associated amino acid sequence or nucleic acid sequence to a cell of said animal.

22. (Original) A method of treating an animal for an imbalance disorder, comprising delivering a therapeutically effective amount of an *atonal*-associated amino acid sequence or nucleic acid sequence to a cell of said animal.

23. (Original) A method of treating an animal for a joint disease comprising delivering a therapeutically effective amount of an *atonal*-associated amino acid sequence or nucleic acid sequence to a cell of said animal.

24. (Original) A method of treating an animal for an abnormal proliferation of cells comprising delivering a therapeutically effective amount of an *atonal*-associated amino acid sequence or nucleic acid sequence to a cell of said animal.

25. (Original) A method of treating an animal for an abnormal proliferation of cells comprising altering *atonal*-associated nucleic acid sequence or amino acid sequence levels in a cell.

26. (Original) A method of treating an animal for a disease that is a result of loss of functional *atonal*-associated nucleic acid or amino acid sequence comprising delivering a therapeutically effective amount of an *atonal*-associated amino acid sequence or nucleic acid sequence to a cell of said animal.

27. (Currently Amended) The method of claim 18, 19, ~~20~~, 21, 22, 23, 24, 25, or 26, wherein said amino acid sequence or nucleic acid sequence is delivered by a delivery vehicle.

28. (Currently Amended) The method of claim 18, 19, ~~20~~, 21, 22, 23, 24, 25, or 26, wherein said amino acid sequence or nucleic acid sequence is delivered by a delivery vehicle, and wherein said delivery vehicle is selected from the group consisting of an adenoviral, vector, a retroviral vector, an adeno-associated viral vector, a plasmid, a liposome, a nucleic acid, a peptide, a lipid, a carbohydrate and a combination thereof.

29. (Currently Amended) The method of claim 18, 19, ~~20~~, 21, 22, 23, 24, 25 or 26, wherein said amino acid sequence or nucleic acid sequence is delivered by a delivery vehicle, wherein said delivery vehicle is selected from the group consisting of a viral vector or a non-viral vector.

30. (Currently Amended) The method of claim 18, 19, ~~20~~, 21, 22, 23, 24, 25 or 26, wherein said amino acid sequence or nucleic acid sequence is delivered by a delivery vehicle, and wherein said delivery vehicle is a cell.

31. (Currently Amended) The method of claim 18, 19, ~~20~~, 21, 22, 23, 24, 25 or 26, wherein said *atonal*-associated amino acid sequence or nucleic acid sequence is Math1.

32. (Currently Amended) The method of claim 18, 19, ~~20~~, 21, 22, 23, 24, 25 or 26, wherein said *atonal*-associated amino acid sequence or nucleic acid sequence is Hath1.

33. (Currently Amended) The method of claim 18, 19, ~~20~~, 21, 22, 23, 24, 25 or 26, wherein said cell contains an alteration in an *atonal*-associated nucleic acid sequence or amino acid sequence.

34. (Currently Amended) The method of claim 18, 19, ~~20~~, 21, 22, 23, 24, 25 or 26, wherein said amino acid sequence has at least about 80% identity to about 20 contiguous amino acid residues of SEQ ID NO:58 (Hath1).

35. (Currently Amended) The method of claim 18, 19, ~~20~~, 21, 22, 23, 24, 25 or 26, wherein said nucleic acid sequence encodes a polypeptide which has at least about 80% identity to about 20 contiguous amino acid residues of SEQ ID:58 (Hath1).

36. (Currently Amended) The method of claim 18, 19, 20, 21, 22, 23, 24, 25 or 26, wherein said cell is a human cell.

37. (Currently Amended) The method of claim ~~20 or~~ 21, wherein said delivery comprises injecting into an inner ear a therapeutically effective amount of an *atonal*-associated amino acid sequence or nucleic acid sequence.

38. (Original) The method of claim 23 wherein said joint disease is osteoarthritis.

39. (Original) The method of claim 24 or 25, wherein said cell is a cancer cell.

40-46. (Cancel)

47. (Original) A fusion protein comprising an *atonal*-associated amino acid sequence or fragment thereof and a desired amino acid sequence.

48. (Currently Amended) A nucleic acid sequence encoding ~~the fusion protein of claim 47~~ a fusion protein, said fusion protein comprising an *atonal*-associated amino acid sequence or fragment thereof and a desired amino acid sequence.

49. (Original) A method of delivering an *atonal*-associated amino acid sequence to an animal, wherein the method comprises administering to the animal a nucleic acid sequence encoding an *atonal*-associated amino acid sequence and a nucleic acid sequence encoding an additional therapeutic amino acid sequence.

50. (Original) The method of claim 20, wherein the method comprises delivering an *atonal*-associated nucleic acid sequence and a second nucleic acid sequence encoding a non-*atonal* associated therapeutic agent.

51. (Original) The method of claim 21, wherein the method comprises delivering an *atonal*-associated nucleic acid sequence and a second nucleic acid sequence encoding a non-*atonal* associated therapeutic agent.

52. (Original) The method of claim 22, wherein the method comprises delivering an *atonal*-associated nucleic acid sequence and a second nucleic acid sequence encoding a non-*atonal* associated therapeutic agent.

53. (Original) The method of claim 24, wherein the method comprises delivering an *atonal*-associated nucleic acid sequence and a second nucleic acid sequence encoding a non-*atonal* associated therapeutic agent.

54. (Original) The method of claim 26, wherein the method comprises delivering an *atonal*-associated nucleic acid sequence and a second nucleic acid sequence encoding a non-*atonal* associated therapeutic agent.

55. (Currently Amended) ~~The composition of claim 40,~~ A composition comprising an *atonal*-associated nucleic acid sequence in combination with a delivery vehicle, wherein said delivery vehicle results in delivery of a therapeutically effective amount of *atonal*-associated nucleic acid sequence into a cell, wherein the composition further comprises an additional ~~amino acid or~~ nucleic acid sequence that is not an *atonal*-associated nucleic acid sequence ~~or amino acid sequence~~.